Original Research Paper

Volume - 14 | Issue - 09 | September - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Medicine

KASABACH MERITT SYNDROME-A RARE PRESENTATION AND MANAGEMENT

Dr Navresham Singh	Junior Resident 2nd Year, Department Of Medicine, Hind Institute Of Medicine Science, Sitapur
Dr Nishant Kanodia	Proff And Hod Of Medicine, Department Of Medicine, Hind Institute Of Medicine Science, Sitapur
Dr Varde JagatKumar HareshKumar	Junior Resident 3nd Year, Department Of Medicine, Hind Institute Of Medicine Science, Sitapur
Dr Amrendra Singh Kushwah	Junior Resident 2nd Year, Department Of Medicine, Hind Institute Of Medicine Science, Sitapur

ABSTRACT The consumptive coagulopathy known as Kasabach-Meritt phenomenon (KMP) is linked to tufted angioma, kaposiform hemangioendothelioma, and vascular malignancies. The Kasabach-Merritt syndrome is typified by severe hypofibrinogenemia, increased fibrin split products, profound thrombocytopenia, and potentially fatal fast tumor development. There may also be indications of severe anemia. Vascular tumors are known to recur and KMP can disappear with timely identification and treatment. The clinical presentation, histology, management, and treatment of KMP in minimally tufted angioma and kaposiform hemangioendothelioma are highlighted in this review. To demonstrate the presentation and our treatment of a patient with KMP, a unique clinical case is presented.

KEYWORDS : Thrombocytopenia, Coagulopathy, Sirolimus, Kaposiform Hemangioendothelioma (KHE), Kasabach-meritt Phenomenon (KMP)

INTRODUCTION

A rare side effect that affects newborns with big vascular tumors is known as the Kasabach-Merritt phenomenon (or syndrome). With a quickly growing vascular mass linked to severe thrombocytopenia and coagulopathy, patients typically show symptoms in the first few months of life. It's a potentially fatal illness.KMP is typified by severe thrombocytopenia and hypofibrinogenemia, as first reported by Kasabach and Merritt[1] in 1940. KMP may include anemia and increased d-dimers. In contrast, thrombocytopenia and low fibrinogen develop related to intralesional entrapment with platelet activation and fibrinogen consumption in vascular tumors, such as KHE with concomitant KMP, which increases the risk of bleeding[2]. KMP was once believed to arise in any commonly occurring infantile hemangioma that was growing quickly, but it was later shown to be connected only to more aggressive vascular tumors.[3][4]

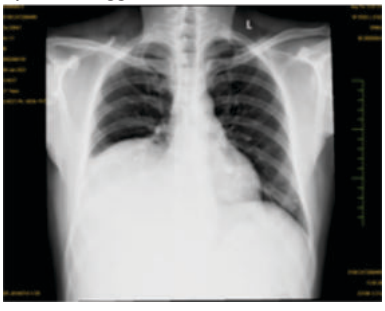

Figure No.1

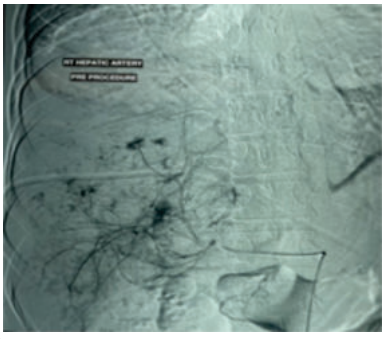

Figure No.2

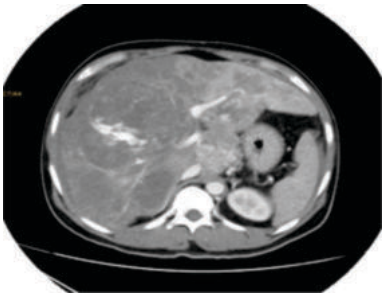

Figure No.3

Case Study

A 32-year-old man arrived with six weeks of RUQ pain and discomfort. The dull, aching pain persisted and became worse with deep breathing movements. He spent ten days with a low-grade fever. He did not exhibit jaundice. Around 2 kg were shed by him at this time. O/E vitals were stable, and the pain visual analog score was 7/10. CVS-average. Reduced air input in the bottom right zone due to RS. P/A: A 5 cm below RCM, palpably sensitive liver was present. The consistency was firm and the surface was smooth. With an overall liver spread of 18 cm, the upper edge of liver dullness was found in the fourth ICS. The current working diagnosis was D/D of HCC and hepatic abscess. USG: Large heterogeneous lesion in right lobe of liver s/o abscess; CXR: elevation of right dome of diaphgram (Figure1).

The largest of the multiple heterogeneous lesions, or > 6 (atypical hemangiomas), on an emergency contrast-enhanced computed tomography (CECT) abdomen (figure3) measured 13.8 x 11.7 x 11.5 cm, with internal high attenuation regions on the non-contrast CT indicating acute hemorrhage.

Hyperdense regions in the arterial phase of contrast-enhanced films indicated active bleeding into the biggest lesion. The bleeding was managed by extremely specific embolization of the right hepatic artery (figure2). The patient recovered without incident. A hematologist suggested that Kasabach Meritt syndrome might be the cause.

Management

- Management of KMS syndrome includes supportive therapy with antifibrinolytics, and plasma (e.g., fresh frozen plasma [FFP]), cryoprecipitate, and platelet transfusions. Other specific therapy

depends on clinical presentation including surgical excision, vascular embolization, compression therapy, immunosuppressive therapy, chemotherapy, and anticoagulation. Interferon therapy should be avoided.

- In the case of thrombosis, intervention should focus on clot resolution. Treatments includes removal of central catheters, thrombectomy, thrombolysis, and anticoagulation. Platelet transfusions may be given as needed to maintain hemostatic parameters. The platelet count may also increase in conjunction with successful anticoagulation.
- For von Willebrand type 2B therapy may include von Willebrand concentrate and combination with platelet transfusions to address hemostasis. While for congenital TTP replacement of ADAMTS-13 can be accomplished through FFP or potentially recombinant ADAMTS-13, currently in clinical trials[9].

DISCUSSION

One type of consumptive coagulopathy caused by platelet aggregation and entrapment within a specific type of hemangioma is known as Kasabach Meritt syndrome [5]. The skin is the most typical location [6], but it can also grow close to important organs, which can cause serious illness [7]. Liu Y et al. reported a case involving a 34-year-old female patient who had a history of multiple giant hepatic hemangiomas during pregnancy and multiple subcutaneous masses that were managed with resection and embolization. The patient also presented with petechia, purpura, multiple subcutaneous masses over her limbs and trunk, distended abdomen, and hepatomegaly. Although the condition is typically diagnosed in infancy [6], there have been adult cases reported. However, at the age of 14, she began to experience skin symptoms.

CONCLUSIONS

With KMP, which has a significant death rate if left untreated, prompt diagnosis and treatment beginning are essential. KMP can resolve and KHE/TA can regress with effective care. Retro peritoneal lesions may have a greater death rate, most likely as a result of a delayed diagnosis and a stronger correlation with KMP. The degree of coagulopathy is substantially correlated with mortality. Long-term consequences include lymphedema, chronic discomfort, and functional impairments, as well as the possibility of recurrence. Based on institutional experience and inclination, we decided to start our patient with KMP and KHE on steroids and sirolimus. After the KMP completely resolved, the steroids were stopped, and the patient is still able to tolerate sirolimus while experiencing full resolution of both KMP and KHE. Vascular tumors are uncommon, and the KMP that goes along with them makes them a big collaborative prospective.

REFERENCES

1. Kasabach HH, Merritt KK. Capillary hemangioma with extensive purpura: report of a case. Am J Dis Child. 1940;59:1063–1070. [Google Scholar]
2. Rodriguez V, Lee A, Witman PM, Anderson PA. Kasabach-Merritt phenomenon: case series and retrospective review of the mayo clinic experience. J Pediatr Hematol Oncol. 2009;31:522–526. [PubMed] [Google Scholar]
3. Sarkar M, Mulliken JB, Kozakewich HP, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with kaposiform hemangioendothelioma and not with common infantile hemangioma. Plast Reconstr Surg. 1997;100:1377–1386. [PubMed] [Google Scholar]
4. Enjolras O, Wassef M, Mazoyer E, et al. Infants with Kasabach-Merritt syndrome do not have "true" hemangiomas. J Pediatr. 1997;130:631–640. [PubMed] [Google Scholar]
5. Osman N. Kasabach – merritt syndrome: a case report. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4949964/#!po=72.2222 [Internet]. PubMed Central (PMC). 2021 [cited 19 October 2021]. Available from:
6. Lewis D., Vaidya R. Kasabach merritt syndrome. https://www.ncbi.nlm.nih.gov/books/NBK519053/ [Internet]. Ncbi.nlm.nih.gov. 2021 [cited 22 October 2021]. Available from:
7. Zou R., Peng F., Yu T., Zeng S., You Y., Chen K., Zou H., Tian X., Zhu C., He X. Kasabach-Merritt Syndrome combined with hypercalcemia: a case report. Exp. Ther. Med. 2017 https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5740515/#!po=47.5000 December. from. [PMC free article] [PubMed] [Google Scholar]
8. Liu Y., Wu X., Ye L., Xu H. Successful treatment of a patient with Kasabach–Merritt syndrome and multiple giant hepatic hemangiomas. J. Int. Med. Res. 2020;48(1) [PMC free article] [PubMed] [Google Scholar]
9. Catherine E. McGuinn MD, ... James B. Bussel MD, in Transfusion Medicine and Hemostasis (Third Edition), 2019